WHAT IS CLAIMED IS:

1.   A method of improving a phenotypic defect in a cell that contains a conformationally defective target protein wherein the conformational defect causes the phenotype defect, comprising contacting a first cell that expresses said conformationally defective target protein with an amount of a protein stabilizing agent that is effective to improve the conformational defect, thereby improving the phenotypic defect of the first cell in comparison with a second cell having the same conformationally defective target protein and phenotypic defect, wherein the second cell is not contacted with a protein stabilizing agent; wherein Congo Red is not the protein stabilizing agent.

2.   A method according to claim 1, wherein the cell is selected from the group of cells consisting of bacterial and eukaryotic cells.

3.   A method according to claim 1, wherein the defective target protein is the gene product of a naturally occurring mutant nucleic acid.

4.   A method according to claim 1, wherein the defective target protein is the gene product of a heterologous nucleic acid.

5.   A method according to claim 1, wherein the defective target protein is selected from the group consisting of the cystic fibrosis transmembrane conductance regulator (CFTR) protein, emphysema and chronic liver disease α-1 anti-trypsin inhibitor, LDL receptor (familial hypercholesterolinemia), β-hexylaminidase (Tay-sachs), fibrillin (Martan syndrome), superoxide dismutase (amyotropic lateral sclerosis), collagen (scurvy) α-ketoacid dehydrogenase complex (maple syrup urine disease), p53 (cancer), type I procollagen pro-α (osteogenesis imperfecta), β-amyloid (Alzheimer's disease), crystallins (cataracts), rhodopsin (retinitis pigmentosa), and insulin receptor (leprechaunism).

6.   A method according to claim 1, wherein the reference protein stabilizing agent is selected form the group consisting of dimethylsulfoxide (DMSO), deuterated water, polyols, sugars, and amino acids and derivatives thereof.

1   7.   The method according to claim 6, wherein the protein stabilizing agent is
2   selected from the group consisting of glycerol, erythritol, trehalose isofluoroside, sorbitol,
3   and polyethylene glycol.

1   8.   The method according to claim 6, wherein the protein stabilizing agent is
2   selected from the group consisting of glycine, alanine, proline, taurine, betaine, octopine,
3   glutamate, sarcosine, gamma-aminobutyric acid, and trimethylamine N-oxide (TMAO).

1   9.   A method according to claim 1, wherein the phenotypic defect is caused
2   by a condition selected from the group consisting of improper folding, improper co- and
3   post-translational modification, improper subcellular targeting, inability to bind biological
4   ligands, aggregation, proteolytic degradation, and any combination thereof.

1   10.   A method according to claim 9, wherein the condition that causes the
2   phenotypic defect occurs in a part of the protein that is selected from the group consisting
3   of pre-sequence, pro-sequence, and mature protein sequence.

1   11.   A screening method for detecting a phenotypically defective cell whose
2   phenotypic defect is due to the presence of a conformationally defective target protein,
3   comprising the steps of
4   contacting a test cell having a phenotypic defect with a protein stabilizing agent,
5   and
6   determining whether such contact is effective to improve the phenotypic defect of
7   the cell.

1   12.   A method according to claim 11, wherein the reference protein stabilizing
2   agent is selected from the group consisting of dimethylsulfoxide (DMSO), deuterated
3   water, polyols, and amino acids and derivatives thereof.

1   13.   A method according to claim 9, wherein the cell is selected from the group
2   of cells consisting of bacterial and eukaryotic cells, in particular yeast, insect and
3   mammalian cells.

1   14.   A method according to claim 11, wherein the defective target protein is the
2   gene product of a heterologous nucleic acid.

15. A method according to claim 11, wherein the defective target protein is selected from the group wherein the defective target protein is selected from the group consisting of the cystic fibrosis transmembrane conductance regulator (CFTR) protein, emphysema and chronic liver disease α-1 anti-trypsin inhibitor, LDL receptor (familial hypercholesterolinemia), β-hexylaminidase (Tay-sachs), fibrillin (Martan syndrome), superoxide dismutase (amyotropic lateral sclerosis), collagen (scurvy) α-ketoacid dehydrogenase complex (maple syrup urine disease), p53 (cancer), type I procollagen pro-α (osteogenesis imperfecta), β-amyloid (Alzheimer's disease), crystallins (cataracts), rhodopsin (retinitis pigmentosa), and insulin receptor (leprechaunism).

16. A method of detecting the relative proportions of PrP^C and PrP^{Sc} present in a composition, comprising:

mixing a composition that comprises prion proteins with a solution wherein only one form, either PrP^C or PrP^{Sc}, is insoluble;

separating the form of PrP that is soluble from the form that is insoluble; and

determining the relative amounts of soluble and insoluble PrP.

17. A method according to claim 16, wherein the PrP is mixed with a solution comprising about 1% Triton X-100 and about 1% DOC at 4 C.

18. A method according to claim 16, wherein the soluble and insoluble forms of PrP are separated by centrifugation.

19. The use of a protein stabilizing agent to improve a phenotypic defect in a cell that contains a conformationally defective target protein wherein the conformational defect causes the phenotype defect, wherein the protein stabilizing agent is selected from the group consisting of dimethylsulfoxide (DMSO), deuterated water, polyols; and amino acids and derivatives thereof.

20. A use according to claim 19, wherein the polyol is selected from the group consisting of glycerol, erythritol, trehalose isofluoroside; polyethylene glycol; and sorbitol.

21. A use according to claim 20, wherein the amino acid or derivative thereof is selected form the group consisting of glycine, alanine, proline, taurine, betaine,

3   octopine, glutamate, sarcosine, gamma-aminobutyric acid, and trimethylamine N-oxide

4   (TMAO).

1   22.   A use according to claim 19, wherein the defective target protein is

2   selected from the group consisting of the cystic fibrosis transmembrane conductance

3   regulator (CFTR) protein, emphysema and chronic liver disease α-1 anti-trypsin inhibitor,

4   LDL receptor (familial hypercholesterolinemia), β-hexylaminidase (Tay-sachs), fibrillin

5   (Martan syndrome), superoxide dismutase (amyotropic lateral sclerosis), collagen

6   (scurvy), α-ketoacid dehydrogenase complex (maple syrup urine disease), p53 (cancer),

7   type I procollagen pro-α (osteogenesis imperfecta), β-amyloid (Alzheimer's disease),

8   crystallins (cataracts), rhodopsin (retinitis pigmentosa), and insulin receptor

9   (leprechaunism).